pathogens are wild strains acquired in the community rather than nosocomial and multiresistant. Based on this, a medico-psycho-social project was initiated, aimed to minimize the time patients have to stay at hospital and to provide the necessary support for successful treatment and life at home. This model is delineated. Though still in lively development, many patients already attend social events, their normal school etc. even during phases of very intensive treatment. Our experience is, that this is very important for all patients with chronic disease. The improvement of quality of life and its effect on the course of the disease will be discussed.

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IS POSSILE TO REDUCE THE TREATMENT OF LYMPHOCYTIC PREDOMINANCE HODGKIN'S DISEASE (LPHD) IN CHILDREN ?

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Purpose: To present the results of treatment of LPHD in children and to discuss the possibility to reduce the intensity of the treatment comparing two periods of therapeutic approach on the same Institution

Material and Methods: From january/80 to december/95, twenty-eight children with PLHD were treated on Pediatric and Radiation Therapy Departments of A C Camargo Hospital - São Paulo - Brazil. This type of histology represented 20.8% of all HD on childhood. The median age was 9 years with male predominance (3:1). The staging system showed 10 stage I, 10 stage II, 6 stage III and 2 stage IV. All patients treated until 1989 (15 children) received chemotherapy (MOPP/ABVD) and involved field radiotherapy (IFRT) at 20Gy. After 1990 the chemotherapy was changed (OPPA/VEEP) and IFRT was done in nine patients and was not done in four (13 treated children)

Results: 26/28 children (92,85%) are alive with local control of the disease and low incidence of side effects. One patient is lost of follow-up and another one is dead. All patients treated after 1990 are alive with a median follow-up of 38 months and lower intensity of side effects treatment. The only patient dead was treated on first period with IFRT and chemotherapy.

Conclusions: The data show that PLHD in children is a high curable diasease. Efforts may de done to reduce de intensity of the treatment increasing the therapeutic results with low incidence.

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NEURORADIOLOGICAL RESPONSE PATTERN OF CHILHOOD LOW GRADE ASTROCYTOMAS (LGA) TO RADIATION THERAPY (RT).

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Results of present chemotherapy (CT) protocols on childhood LGA are often expressed in terms of tumour volume changes (TVC). However, these criteria seem to be of questionable value for these tumours. Therefore, LGA response in terms of TVC to RT, a treatment modality considered potentially curative, has been evaluated and correlated with patient outcome. This study population consisted of 11 of the 37 children enrolled in SIOP clinical trial on LGA, treated with RT from August 1992 and February 1996. RT was delivered to the tumour bed plus margins at a total dose of 54 Gy in 6 weeks. 6 pts were treated at diagnosis for diencephalic syndrome or decreased visual acuity, 3 at disease progression and 2 after CT due to relapsed. After RT, all children were submitted to MRI every 3 months. At present all pts are alive. At a median follow up of 42 months (12-54), 3 children present radiologically stable disease after a partial response in dimensions and contrast enhancement and 8 have stable disease with no change in dimension and very small variation in contrast enhancement. Histological subtype, site of tumor, size at diagnosis and NF1 did not affect response. No differences were observed between pts submitted to RT alone and children previously treated with CT. Contrary to that generally reported in oncology, TVC other than tumour progression do not seem to be reliable criteria to predict patient outcome. A longer follow up is however needed.
Partially supported by Italian Association Against Cancer.

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PROPHYLAXIS FOR CHEMOTHERAPY-INDUCED EMESIS IN CHILDREN WITH CANCER: A COST-SAVING MODEL FOR DEVELOPING COUNTRIES

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The objective of the study was the analysis of efficacy and safety of a cost saving anti-emetic therapy. Thirty seven children with malignant diseases aged 1-17 years, receiving chemotherapy (161 cycles), were treated with 5-HT_3-antagonist ondansetron or metoclopramide. These receiving highly emetogenic therapy (except cisplatin) were given ondansetron in a single dose (i.v. or p.o.) and those given cisplatin were treated with ondansetron in divided doses (3/day). Metoclopramide (0.5-1 mg/kg/dose) was used as an antiemetic agent in a group of children receiving moderately emetogenic chemotherapy. A complete or major response (less than 3 vomiting episodes) was achieved in 90% of all cycles. Ondansetron in a single dose was effective in 98% of cycles of highly emetogenic therapy and was effective in divided doses in 2 patients receiving cisplatin. Metoclopramide was successful in the control of vomiting in patients receiving moderately emetogenic therapy in 94% of cases. In case of failure, an extra dose of ondansetron or ondansetron + dexamethason was given. Eventually, ondansetron given in a single-dose happened to be an efficient antiemetic drug. Metoclopramide used in control of vomiting in moderately emetogenic therapy was highly effective and non-toxic drug.

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IMPLANTABLE VENOUS ACCESS SYSTEM FOR THERAPY IN CHILDREN WITH MALIGNANT DISEASES-ANALYSIS OF COMPLICATIONS.

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The use of pernament intravenous access devices increases comfort of therapy and has become widespread in children with cancers. Therefore, awareness of potential complications is important.

Objectives. The aim of this study was to determine the incidence and clinical characteristics of complications related to long-term vein catheterisation.
Patients and Methods. Data were collected from July 1993 through February 1997. The cohort consisted of 35 pediatric oncology patients who had totally implantable venous access systems /Vascuport, Viggo AB./ The range of age patients was 5 months to 16 yrs 7 months/mean 8 yrs 1 mth/ Individual catheters were in place for a median 516 days /3-1220/, with a total experience of 18. 087 days.
Results. Seven patients died from their disorders prior to completion of the study. Nine vascuport systems had to be removed before completion of scheduled therapy; 4-because of local complications; 2-because of sepsis and 3 because of thrombotic complications. No catheter related death occured. The total incidence of complications leading to removing of systems in our material was 25,7%. The development of right atrial thrombus in one of our patients prompted us to perform echocardiography in 20 patients having central vein catheter as a screen for thrombotic complications.
Conclusions. According to our results routine echocardiographic screening of symptom-free patients is not rewarding, although it is the method of choice for imaging intracardiac thrombus and to confirm diagnosis in symptomatic patients. The long-term use of totally implantable venous access systems in children treated for malignant disease is associated with acceptable complications and offers many advantages in clinical care.

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IMMUNE RESPONSE TO INFLUENZA VACCINATION IN PREVIOUSLY VACCINATED CHILDREN WITH ACUTE LYMPHOBLASTIC LEUKEMIA.

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The aim of the present study was to estimate the immune response to influenza vaccination in children with acute lymphoblastic leukemia who had previously been vaccinated in the epidemic season 1993/94.

Twenty-seven children with leukemia aged 7-16 were immunized in autumn 1996 with a single dose of subunit trivalent purified influenza vaccine ('Influvac', Solphay Duphar) in the Paediatric Dept. of Haematology and Oncology, Medical Academy, Warsaw. The vaccine used in the study consisted of 15 µg of haemagglutinin in 0.5 ml doses of each of the following influenza strains: A/Singapore/6/86 (H1N1), A/Wuhan/395/95 (H3N2) and B/Beijing/184/93 (HB). Antibody production was determined before vaccination and three weeks after vaccination by the haemagglutinin inhibition test (HI). The same parameters were determined for the control group (30 children).

Before vaccination the geometric mean titre (GMT) of antihaemagglutinin antibodies was nearly on the same level for all three influenza strains and ranged between 2.2 and 2.7. After vaccination the GMT values were 37.0 for haemagglutinin H1, 48.8 for haemagglutinin H3 and 82.1 for haemagglutinin HB. The highest mean fold increase of haemagglutinin antibody titres after vaccination which amounted to 30.4 was observed for haemagglutinin HB. For other types of haemagglutinins (H1 and H3) the mean fold increase reached nearly the same level (16.8 and 18.8). In the control group GMT for all anigens was on a low level during the study. Mean fold increase in this group ranged from 1.3 to 2.5.

In conclusion we would like to emphasize that the results point to a significant seroconversion for haemagglutinin after vaccination in comparison with the control group. In the group of vaccinated children we did not observe any infections with the influenza virus. The clinical data we obtained clearly suggest that during an influenza epidemic, children with leukemia in remisssion can be protected by prompt immunization.
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COMPARISON OF CHLORPROMAZINE PLUS DEXAMETHASONE VS ONDANSETRON VS TROPISETRON IN THE TREATMENT OF EMESIS INDUCED BY HIGHLY AND MODERATELY EMETOGENIC CHEMOTHERAPY IN PEDIATRIC PATIENTS WITH MALIGNANCIES

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OBJECTIVES. To compare the efficacy and safety of two 5-HT_3 receptors antagonist, ondansetron (OND) and tropisetron (TRO), versus chlorpromazine (CHL) + dexamethasone (DEX) in children receiving highly and moderately emetogenic chemotherapy. **PATIENTS AND METHODS.** Children with solid malignant tumors who were chemotherapy-naive, were randomized in a double-blind, placebo, cross-over trial in order to be administered CHL 0,3 mg/kg and DEX 2 mg/m^2 intravenously (i.v.) 30 min before and 6 and 12h after chemotherapy or OND 5 mg/m^2 i.v. 30 min before and 12h after chemotherapy or TRO 0,2 mg/kg i.v. 30 min before and 12h after chemotherapy as antiemetic therapy. **RESULTS.** 46 patients for a total number of 302 chemotherapy cycles were evaluated. *Highly emetogenic chemotherapy*: A complete response (0 emetic episodes) for emetic episodes was achieved in 34% and 28,1% of the patients in OND and TRO groups respectively, versus 16,7% in CHL + DEX group ($p < 0.01$). A major response (1-2 emetic episodes/day) for emetic episodes was achieved in 38,3% and 31,2% of the patients in OND and TRO groups respectively, versus 14,6% in CHL + DEX group ($p < 0.01$). There was no statisticaly significant differences between OND and TRO groups. *Moderately emetogenic chemotherapy*: Results did not show any significant differences between the three antiemetics regimens. *Adverse events*: Somnolence was more frequent in patients submitted to CHL + DEX regimen (57,8%) than in OND (16,5%) and TRO (7,5%) groups ($p < 0.001$). **CONCLUSIONS.** OND and TRO regimens were superior to CHL + DEX in children receiving highly emetogenic chemotherapy. Efficacy of the three antiemetics regimen was similar in moderately emetogenic chemotherapy

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TROPISETRON AS PROPHYLAXIS FOR CHEMOTHERAPY INDUCED EMESIS IN CHILDREN WITH SOLID TUMORS

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Nausea and vomiting are the most distressing side effects associated with the administration of cytotoxic chemotherapy. This study aims to assess the efficacy of a 5HT3 receptor antagonist tropisetron, in the control of emesis and nausea in children with malignant solid tumors receiving a variety of highly emetogenic chemotherapy (CT) combinations.

Seventy-nine children with a median age of 9 years (range, 6 months-18 years) were administered tropisetron (Navoban-Sandoz), dosed once daily at 0.2 mg/kg (with a maximum of 5 mg daily) i.v. immediately before CT for a total of 371 CT courses. On the first 5 days of each course of CT, emetic response to Navoban for 24-hour period was graded as complete (no emetic episode), partial (1-5 emetic episode) or failure (>5 emetic episode). Nausea was graded as none, mild (feeling sick), moderate or severe (feeling very sick). In the first 24 hour period after starting CT complete response regarding emesis (acute emesis) was obtained in 58%, partial response in 36% of all courses. Nausea was graded as none in 64% mild or moderate in 31% and severe in 5% of all courses. In the "worst day" of treatment, complete response was obtained in 45% and partial response in 46% of all courses, nausea was recorded as none in 58%, mild or moderate in 36% and severe in 6%. In 50/79 chemotherapy naive patients, acute emesis response in the first course of CT was graded as complete in 58%, partial in 34% and as failure in 8%. No nausea was seen in 50%; mild

nausea was seen in 46% and severe in 4%. The second and subsequent courses yielded similar percentages. Mild side effects were documented in 12/371 (3.2 %) courses.

The results of this study suggest that tropisetron is a safe, well tolerated and effective anti-emetic in the treatment of children receiving highly emetogenic chemotherapy.

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C REACTIVE PROTEIN (CRP) AND FIBRINOGEN ARE 2 EARLY MARKERS TO ASSESS RESPONSE TO EMPIRIC ANTIBIOTHERAPY IN FEBRILE NEUTROPENIC CHILDREN.

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Objectives of the study : in a prospective study, we evaluate 3 usual biological markers of inflammation to assess the response to empiric antibiotherapy in febrile neutropenic children.

Material and methods : from jan. 95 to feb. 96, all febrile neutropenic children followed for cancer in our unit, were enrolled in a randomised study of 2 antibiotics associations (Group A : continuous infusion of Ceftazidime and Vancomycine and once daily administration of Amikacine. Group B : once daily administration of Ceftriaxone, Teicoplanine and Amikacine). Dosage of CRP, fibrinogen and orosomucoid were realised at H0, H24 and H48 of the treatment. Success was considered when apyrexia was obtained and remained stable after 72 h of treatment. Informed consent from the parents was obtained and the study was approved by the ethical comity of our hospital.

Results : 76 of 84 febrile neutropenic episods were evaluable : 39 in the group A and 37 in the group B. Both groups were comparable for age and type of diseases. Orosomucoid was tested only in 12 pts.

		CRP (mg/l) Mean (±SD)		Fibrinogen (g/l) Mean (±SD)	
		H0	H48	H0	H48
Group A	Success (n=23)	61,83 (79,72)	24,35 (23,84)	4,96 (1,34)	4,18 (1,16)
	p*	NS	<0,001	NS	<0,001
	Failure (n=16)	70,75 (121,72)	127,75 (108,62)	4,85 (1,43)	6,89 (1,27)
Group B	Success (n=20)	55,47 (68,63)	27,53 (44,98)	4,87 (1,34) (n=15)	3,95 (1,07)
	p*	NS	0,002	NS	<0,001
	Failure (n=17)	70,88 (72,92)	103,59 (86,38)	4,89 (1,61) (n=11)	7,24 (1,01)

* t Student's Test. NS : Non significative

Conclusion :CRP and fibrinogen are 2 early markers to appreciate response to empiric antibiotherapy in febrile neutropenic children.

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THE MANAGEMENT OF FEBRILE NEUTROPENIC EPISODES IN CHILDREN WITH CANCER

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Management of children with cancer with more intensive chemotherapy almost always results in fever with severe neutropenia. Success of management of these episodes depend on the causative organism, the severity of neutropenia and the efficiency of the antibiotic used.Fifty three neutropenic child with cancer, with neutrophil counts less than 500 cells/mm and fever >38.5 C for more than 24hours, were assessed in our pediatric oncology unit. Common sites of infection were the respiratory tract followed by the GI tract and the urinary tract.Forty five positive cultures were obtained during those episodes. They were 27 positive blood cultures, 10 positive throat swabs and 8 positive urine cultures. Gram -negative bacteria comprised (63%) of the isolates. The most common gram-negative organisms were Klepsiella Oxytoca (24%) and Citrobacter Freundii (10%). Gram - positive organisms were seen with lesser frequency and represented (37%) of isolates. Coagulase negative staphylococcus was the predominant gram- positive organism (13%). Fungal infection (Candida albicans) was identified during 4 febrile neutropenic episodes. Our data clearly shows the predominance of gram- negative bacteria over gram positive in our pediatric oncology unit. Early initiation of empiric antibiotic regimens is crucial in effective management of these cases.

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CHRONIC DISSEMINATED CANDIDIASIS: DO WE ACHIEVE A BETTER OUTCOME TODAY?

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Chronic disseminated candidiasis (CDC) is an infrequent complication in neutropenic patients. It has, however, been associated with significant morbidity and mortality as high as 50% in the past years. We reviewed our recent experience on CDC which includes 4 cases treated in the last 3 years in our department. The patients were 3 boys (A, B and D) and 1 girl (C) of ages 5-13 years. Patients A and B suffered from acute lymphoblastic leukemia, C from acute myeloblastic leukemia and D from a B-cell non Hodgkin's lymphoma. All 4 children were receiving intensive antineoplastic chemotherapy and the disease occurred during profound myelosuppression after an average of 4 neutropenic cycles. Fever was the main symptom of infection lasting for approximately 12 days. Patient D suffered from preceding eosophagitis. Typical lesions were shown by ultrasonography and computed tomography: in A and C, in the liver only; in B, in the liver, spleen and right kidney; and in D, in the liver and spleen. During the progress of CDC, neutrophil count ranged between 600 and 9,500/μl. In addition, alkaline phosphatase was elevated in 3 patients. Repetitive blood cultures drawn before and during CDC episodes were negative. In patients A, B and D, *C. albicans* grew from pharyngeal cultures before antifungal therapy. Due to therapy-related profound thrombocytopenia, liver biopsy was not possible in any of these patients. All 4 patients received liposomal amphotericin B, 5 mg/kg/day for 180 (A and D), 90 (C) and 150 (B) days. B and C also received fluconazole, 5mg/kg/day, for an additional 30 days. B, C and D exhibited clinical and imaging recovery. They were then treated with further antineoplastic chemotherapy without relapses of the infection. Patient A improved but succumbed to a relapse of his underlying malignancy. With early diagnosis, even presumptively in the absence of better diagnostic tools, and prompt initiation of antifungal therapy, CDC appears to have improved outcome compared to previous reports.

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SUCCESSFUL CARBOXYPEPTIDASE RESCUE IN A CHILD WITH RENAL FAILURE AFTER HIGH DOSE METHOTREXATE ADMINISTRATION.

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OBJECTIVE: High Dose Methotrexate (HD MTX) prolonged intravenous infusions are generally administered with adequate fluid hydration and alkalinization and folinic acid rescue. These preventive measures are generally adequate to prevent MTX related toxicity. Since MTX is primarily

excreted by kidneys, the onset of renal failure during HD-MTX can however represent a life-threatening complication. The successful Carboxypeptidase (CPD) rescue in a child with renal failure after HD MTX administration is here reported. CASE REPORT: B.A., female, was diagnosed with acute lymphoblastic leukemia (ALL) at the age of 14 years and was treated according to AIEOP ALL 9102 protocol. After Induction therapy complete remission was obtained; HD MTX (5 gr/sqm/24 hrs infusion i.v.) was given as Consolidation therapy. During the first course of HD MTX administration the patient experienced several episodes of nausea and vomiting despite i.v. administration of ondansetron. At the end of HD MTX infusion (24th hour) plasma levels of the drug were 192 mmol/L and a marked reduction of urine output was observed. Weight and creatinine values rapidly increased. Hydration and alkalynization were continued, furosemide was administered to maintain adequate diuresis and folinic acid was given at the dose of 2500 mg/sqm q 6 hrs. Between the 36th and the 48th hour two hemodialysis courses were performed. At the 48th hour MTX plasma level was 19 mmol/L. Because of expected severe MTX related toxicity, CPD was obtained for compassionate use and administered at the dose of 50/IU/Kg i.v. at hours 72 and 76 after HD MTX start without adverse reactions. MTX plasma level measured by HPLC was 1.0 mmol/L 1 hour after the firt dose of CPD and 0.5 mmol/L after the second one. Conversely MTX plasma level measured by fluorescence polarization immunoassay was consistently higher. Renal function recovered within few days. Folinic acid dosage was progressively decreased and discontinued after 1 week. No toxicity on liver, mucosae or bone marrow was observed. CONCLUSIONS: In this experience the use of CPD has been proven to be effective in preventing life-threatening MTX related toxicity and is thus recommended for patients with renal failure. MTX plasma levels, investigated by traditional fluorescence polarization immunoassay, could be overestimated due to cross reactivity with inactive MTX metabolites. In this case a reliable evaluation of the MTX plasma levels can only be obtained by HPLC.

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RENAL CELL CARCINOMA IN CHILDREN. SINGLE INSTITUTION EXPERIENCE.

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Renal cell carcinoma (RCC) is rare in childhood, however may happen also in that age and than surprises the treating team. Aim of the study was to evaluate pathology, staging, treatment and outcome in pts treated for RCC in our department. Patients. Six pts with RCC were treated in our institution (1986-1995). Average age at diagnosis was 10 years and 4 months, detailed pathology review evidenced CCR with mixed tubular and papillary patterns in 4 pts. CCR with clear and eosinophillic cells in 1 and pure papillary eosinophyllic type in 1. Three RCC were classified as G4, 1 as G3 and 2 as G2. Staging: 2 pts (both G2) were staged II and 4 - were staged III. One RCC developed in horseshoe kidney. Treatment: 5 pts received preoperative chemotherapy as they were considered to be Wilms' tumours (4-week VCR/ActD); only minimal response had been observed. Radical nefrectomy and regional lymph nodes dissection were performed in all cases. In 4 of 6, lymph nodes were invaded histologically. Four pts received postoperative chemotherapy and RTX, 2 (more recent cases) were treated with IL-2 and IFN-alfa. Results and outcome: 2 of 6 pts are alive and disease free for more than 3 years: both were stage II G2 pts, one received IL-2 and INF-alfa. Four stage III pts died of metastatic relapses to lungs and liver. Summary: Experience gained in management of RCC in children is weak, however, adjuvant treatment with IL-2 and INF-alfa, currently used in adults seems promising also in children. Surgery should be more extensive than that adviced in Wilms' tumour and radical lymph nodes dissection should be considered.

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COMBINED RESECTION OF THE ABDOMINAL AORTA AND THE INFERIOR VENA CAVA FOR RETROPERITONEAL RHABDOMYOSARCOMA INVADING THE AORTIC BIFURCATION

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The prognosis of rhabdomyosarcoma has been improved with multimodal therapy, but it is still poor when only an incomplete resection was performed. We report a rare case of retroperitoneal rhabdomyosarcoma at the aortoiliac bifurcation, which was successfully resected en bloc with combined resection of the abdominal aorta and the inferior vena cava (IVC). To our knowledge, this is the first case in which aortic bypass graft was replaced for retroperitoneal tumor in childhood.

The patient was a 11-year-old boy with an abdominal tumor, measuring 8x10 cm. Biopsy revealed rhabdomyosarcoma of alveolar type and imaging techniques showed a nonmetastatic retroperitoneal tumor which involved the abdominal aorta, IVC and common iliac vessels. Since the tumor was not sensitive to combined chemotherapy, but was slightly sensitive to irradiation, we decided to perform tumor resection and intraoperative irradiation. At operation the tumor densely invaded to the abdominal aorta and IVC so that combined resection of the major vessels were performed, indicating IRS-group 2 disease (grossly resected tumor with microscopic residual disease). The infrarenal abdominal aorta, 8 mm in diameter, and iliac arteries were replaced with a 12x6 mm Y shaped Dacron graft. Prosthetic replacement of the IVC was not performed. After aortic replacement, intraoperative irradiation (15 Gy) was made. Histologic study revealed tumor invasion to adventitia of the aortic wall. At 12 months follow up, there is no tumor recurrence with patency of the graft on MR angiography.

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A CASE OF METACHRONOUS MULTIPLE COLORECTAL CANCER WITH GENOMIC INSTABILITY IN A CHILD

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BACKGROUND
DNA-replication errors (RERs) have been closely linked to hereditary non-polyposis colorectal cancer (HNPCC) syndrome. Ubiquitous changes in the length of simple DNA sequences between constitutional and tumor DNA occur in about 90% of cases with HNPCC in adults. In cases with pediatric colorectal cancers, however, the incidence of such microsatellite instability has not been documented in detail. The analysis of microsatellite instability was performed on a case of metachronous multiple colorectal cancer in a child.
CASE REPORT
A 12-year-old male was treated surgically because of the synchronous multiple cancers in the sigmoid and the rectosigmoid colon. At the age of 23, the synchronous multiple cancers were again presented in the residual transverse colon and the cecum. Subtotal colectomy was performed. P53 gene mutation was not detected in both metachronous tumors. As no obvious family history of the malignant neoplasms was obtained, this case did not meet the minimum criteria of HNPCC. To clarify the patient's genomic disorders, two microsatellite loci (D2S123 and D3S1067) were studied using PCR method. RER was positive in both of two loci, which implies that this patient is a possible initiator of HNPCC family. He has been followed up intensively to date not to overlook further multiple organ neoplasms.
CONCLUSION
The analysis of RERs in pediatric patients with the lack of HNPCC characteristics, is supposed to be useful to detect the high risk group of multiple organ cancers.

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A REPORT OF 61 CHILDREN WITH MEDULLOBLASTOMA

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The medulloblastoma is a tumor of the posterior fossa, standard treatment consists of surgical resection followed by radiation to the craniospinal axis and chemotherapy.
We report here a series of 61 medulloblastoma treated at our institution between 01/05/82 and 31/12/94. The mean age of the patients was 7 years at diagnosis (from 6 months to 15,8 years) with a sex ratio of 1,8.
The site of the tumor was found to be central in 48 cases and lateral in 13 cases. Surgical results were : 40 complete or gross total resection, 14 subtotal and 7 partial resection.
59% had a medulloblastoma with standard risk (complete or gross total resection and no brain and neuroaxis dissemination by imagery and cerebrospinal fluid cytologic examination) and 41% had a bad risk.
16/61 received only radiotherapy and 45/61 an association of chemotherapy and radiotherapy after surgery.
52% had a relapse, 27 before 3 years after the end of the treatment.
All these patients died of their tumor except one who survived of recurrences ranging from one to 30 months (median 6 months).
The median follow-up time is 69 months and the 5 years overall survival rate is 52%.
The most important risk factors are the quality of surgical exerese and absence of CNS metastasis.

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PVB therapy and complete resection of tumour improved the prognosis of yolk sac tumour

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Yolk sac tumour is the most predominant tumour in the teratomatous malignancy in the childhood and progress aggressively. We experienced eleven cases in which seven cases (64%) followed good prognosis. Onset age was 8 months-15 years old.
Primary sites were sacrococcigeal 3, testis 2, ovary 2, retroperitoneal 2, mediastinum 1, abdominal wall 1. Stage I in 2, II in 2, III in 4 and IV in 3. AFP value was 1600-290000 ng/ml, LDH 218-1479 IU/l. All primary tumours were surgically extirpated: four huge masses were first treated with PVB therapy (CDDP 75 mg/m^2, Bleomycin 10 mg/m^2,Vinblastin 7 mg/m^2) following biopsy, then resected surgically. All stage IV died of lung metastasis, but other stages I-III except one stage II survived. This exceptional stage II died of intra-abdominal recurrence because of tumour cell spilled out during operation.
In summary, PVB therapy controls yolk sac tumour very well and makes surgical resection of tumour more safe and complete. This combination therapy improves the prognosis of yolk sac tumour. However stage IV and intraoperative-tumour spilled out still remain incurable.

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TWO SPORADIC CASES OF BANNAYAN-RILEY-RUVALCAVA SYNDROME (BRRS)

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Objective: BRRS is a rare disorder characterized by macrocephaly and multiple soft tissue and visceral hamartomas. This report presents clinicopathologic findings of two sporadic BRRS cases with macrocephaly, lipomas, hemangiomas, and lymphangiomas.
Case reports: Case 1 is a girl who had developed multiple subcutaneous lipomas on the chest and the abdominal wall at the age of 5 months. CT scan showed macrocephaly with slight ventricular dilatation and cervical, mediastinal, retroperitoneal and abdominal wall lipomas. The lipomas were extirpated 3 times by the age of 34 months. Hemangiomas on the left arm and right upper mediastinum and multiple cutaneous lymphangiomas on the chest appeared at age of 3 years. At the age of 5 years she developed cardiac failure due to an enlargement of the cervicomediastinal arteriovenous fistulous hemangiomas. Excsion, irradiation and embolizations did not relief from cardiac failure and she died of cardiac and respiratory failure and septic shock at the age of 9 years. Case 2 is a girl who had developed right ovarian granulosa cell tumor at the age of 3 months and multiple lipomas at the age of 1 year. Macrocephaly, multiple cutaneous lymphangiomas, cutaneous and visceral hemangiomas, and lipomas developed with growth. Now she is 7 years old who has been needing multiple excisional operations.
Conclusions: BRRS is a still uncertain clinical entity and needs further pathogenetic investigations and it is required more sophisticated clinical treatment strategy.

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STROMAL TUMORS OF THE GASTROINTESTINAL TRACT IN CHILDREN. THREE CASES WITH PROLONGED FOLLOW-UP.

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Stromal tumors of the gastrointestinal tract (Leiomyoma, leiomyoblastoma and leiomyosarcoma) are very uncommon in children. We describe here three such tumors.

Case 1- A 10-year-old girl presented with hematemesis and melena. Upper gastrointestinal series and arteriography revealed a gastric tumor. Sub-total

gastrectomy was performed. Histologic study showed multiple **epithelioid leiomyomas** (leiomyoblastomas). Resection margins were free of tumor. Local recurences occured 11 and 18 years later and surgical resections were performed. Twenty-four years later a multifocal recurence occured (esophagal and pulmonary lesions).

Case 2- A 10-year-old boy underwent urgent laparotomy for massive hematemesis. A tumor of the gastric antrum was seen and sub-total gastrectomy was performed. Histologic study showed an **epithelioid leiomyoma**. Resection margins were free of tumor. Local recurrence was resected 7 years later. Sixteen years later local recurrence occured with a solitary pulmonary nodule. Both lesions were resected. Histologic study showed a gastric **epithelioid leiomyosarcoma** with **pulmonary chondroma**. Investigations for functioning extra-adrenal paraganglioma was negative (incomplete **Carney's Triad**). The patient is alive with no evidence of disease seven years later.

Case 3- A 10-year-old girl was admitted for a mass of the left flank. Rectosigmoidoscopy and CT scan showed a tumor of the sigmoid colon. Surgical resection and histologic study of the tumor disclosed a **low grade leiomyosarcoma**. Resection margins were free of tumor. Adjuvant chemotherapy was performed (Four cycles with Ifosfamide, Vincristine, Actinomycine). The patient is alive with no evidence of disease seven years later.

These reports show the need for prolonged follow-up of patients with gastrointestinal stromal tumors, precise prognosis being difficult to predict. Late recurrences may occur despite complete surgical resection. Careful histologic examination is necessary to correctly classify these tumors, and to identify malignant potential. The role of chemotherapy and radiotherapy is not well defined for children. **Collaborative multicentric studies are necessary to elaborate efficient therapy for children with gastrointestinal stromal tumors.**